

Dercum (F. X.)

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ON THE SIGNIFICANCE OF OPTIC NEURITIS, BLINDNESS, DEAFNESS AND THE KNEE- JERK IN CEREBELLAR DISEASE.¹

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AS is well known, diseases of the cerebellum present some of the most recondite problems in physiology.

Many of the symptoms are both inconstant and variable. They are difficult of explanation and frequently impossible to correlative. Among the symptoms are muscular incoordination, festination, vertigo, vomiting, headache, nystagmus, optic neuritis, difficulty of articulation, polyuria, anomalies of the knee-jerk, blindness, muscular weakness and convulsions. I desire to call attention more especially to the optic neuritis, the blindness, the deafness and the modifications of the knee-jerks.

Before entering into a detailed discussion, it may be well to relate a number of cases in which the above symptoms were present:

CASE I.—H. S., a boy, 5 years of age, was brought to the Jefferson Hospital in January, 1892; father, mother, brother and sister were perfectly healthy. When about one year old he had learned to walk as other children, and talk and play, and seemed in every respect normal. Nothing unusual was noticed in him until about four years of age. The father noticed at this time that he began to stumble while walking, and that this stumbling became more and more marked until, at last, the child could not walk alone. During this time the mental condition remained good.

On examination at the hospital it was found that the child was well nourished, though the head seemed dis-

¹ Read before the meeting of the American Neurological Association, July, 1893.



proportionately large, measuring some $20\frac{5}{8}$ inches in circumference. On standing he swayed slightly forward and backward. When he attempted to walk, he spread his legs widely apart and was evidently excessively ataxic and staggered badly. This ataxia was not limited to the legs, but was also well marked in the hands; for, on asking the child to pick up a small object, as a penny, the movements were very irregular, so as to suggest the "hovering hand" of Freidreich's Disease. It was equally marked on both sides. There was no appreciable weakness in the limbs, and sensation was everywhere normal. The muscles, however, seemed everywhere relaxed. The knee-jerks were absolutely nil. The pupils were rather large, but reacted well to light. There was present also lateral nystagmus marked in both eyes. An examination of the eye ground revealed marked double optic neuritis. The child talked intelligently for its age. There was also a high degree of hypermetropia, but no other peculiarity of vision.

On October 21, 1892, the patient was again examined, and at this time measurement of the head revealed $21\frac{5}{8}$ inches, a decided increase in circumference. The child was at this time totally unable to stand; not from weakness, but from excessive ataxia. The pupils were now extremely dilated and irresponsive. The *knee-jerks had now returned* and were grossly exaggerated; and there was also *total blindness*. Severe epileptiform convulsions had also occurred. A few days later the child was removed from the hospital and passed beyond observation.

CASE II.—S. S., female; white; aged 43; single. (Jefferson Hospital). Family history negative. Mental condition somewhat impaired, so that personal history was elicited with some difficulty. Patient had, however, noticed for a year and a half previously that her gait had become unsteady. Several months later had noticed difficulty with the vision. At the same time headache had set in and vomiting had also recurred repeatedly. Three months ago she became *entirely blind*. Her present condition is as follows: stands with feet widely separated and tends to sway forward and backward with the slightest push. Her equilibrium, when disturbed, is regained with difficulty. If the feet be placed close together she falls. In order to walk she takes short steps with the feet wide apart, swaying and staggering as she moves.

This gait being continued, it is noticed that she walks in a circle always to the right. Occasionally she stops as if to regain her equilibrium. The diameter of the circle is about twelve feet. There is marked incoordination of both arms, as is noticed on her attempt to pick up objects, and in attempting to dress herself. The incoordination is most marked in the left arm. *The knee-jerks are much exaggerated*; most, however, on the left side. Tendo achilles jerk present; no ankle clonus; elbow jerk plus, and much more marked on the left side. The dynamometer registers twenty with the right hand and thirty-five with the left (patient right-handed); no anæsthesia is anywhere detected. Occasionally, however, she fails to locate the impression properly, mistaking at one time the little finger for the thumb, the back of the hand for the palm, and making similar errors in the feet. Recognition of tactile impressions is somewhat delayed, due apparently to an increased cerebral reaction-time. Differentiates hot and cold correctly. Her memory is evidently impaired; thought appears somewhat slow; speech is halting and at times jerky. Articulates at times indistinctly. Complains of occipital and frontal headaches, the occipital being the most severe and most marked in the morning. No local palsies, no vomiting, no tremor, total blindness; pupils dilated and fixed; vertigo, nystagmus and dullness of hearing.

Examination of the eye ground reveals a high grade optic neuritis.

CASE III.²— A. M., white, aged 28, was admitted to the nervous wards of the Philadelphia Hospital, November 12, 1890. He had been in the German Hospital for some five months previous, and had there suffered from headache and pain in the back of the neck. He had also a markedly staggering gait; was subject to epileptiform attacks, during which consciousness was lost and in which rigidity predominated.

Ophthalmoscopic examination disclosed a high grade optic neuritis on either side with impairment of vision. Marked deafness was also noted. These symptoms increased in intensity, until finally total blindness and total deafness ensued. The urine was excessive in quantity, of low specific gravity, and free from both albumen and sugar.

² Previously published, Philadelphia Hospital Reports, Volume I., 1891.

When admitted to the Philadelphia Hospital, he was unable to stand. There was excessive general weakness, absolute loss of sight and hearing, and also, as far as we could determine, loss of smell and taste (?) Cutaneous sensibility, as far as it could be studied under these conditions, appeared to be well preserved. No paralysis of sphincters. The knee-jerks could, with difficulty, be elicited and were practically absent. Communication with patient was only possible by writing with the index finger upon the palm of his hands. Under these difficulties we learned again of his headache, that it was agonizing, and, if possible, steadily growing worse. The epileptiform attacks were also repeated. In these his arms and legs became very rigid, and to the rigidity would be added a rapid clonic movement of small extent.

An ophthalmoscopic examination by Dr. de Schweinitz, revealed marked optic neuritis, dilated, immovable pupils, and nystagmus; also slight divergent strabismus due to loss of fixation. Blindness and deafness were absolute.

Owing to the fact that the pain in the head was markedly increased by percussion of the left frontal region, it was deemed, after consultation with my colleagues, expedient to perform an exploratory operation in this region. This was done by Dr. Hearn. No noteworthy feature presented itself in the trephine opening, except that the brain bulged greatly; and further, that it fluctuated markedly upon palpation. Exploratory puncture was, after a brief consultation, decided upon, with the result that between five and six ounces of fluid escaped from the left lateral ventricle. The operation was terminated at this point. A few hours later the patient rallied, and was evidently much relieved as regards the headache. However, on the third day hernia cerebri began to make its appearance, and concomitantly there set in gradually coma, and finally death on the fifth day.

The autopsy revealed, among other changes of minor importance, a tumor of the cerebellum. On carefully separating the quadrigeminal bodies from the cerebellum, a large jelly-like mass, the size of a pigeon's-egg, was disclosed, occupying the central lobe. The peripheral portion of the tumor was excessively soft, fairly attached to and merged with the white substance of the right cerebellar hemisphere. It appeared to have originally sprung from this region. It was simply in contact with the white substance of the left hemisphere, having hol-

lowed out a space for itself in these structures by pressure and absorption. The roof of the fourth ventricle and the superior peduncles of the cerebellum had been partially destroyed by the growth.

CASE IV.—E. M., a boy, aged 5 years and 9 months, presented himself at the Jefferson Hospital on July 5, 1893. Father and mother are living and well. Three brothers and one sister, all older, are likewise well.

When six months old he was quite ill from "teething and summer complaint." Made, however, a good recovery, and was in good health from this time on until March 20th of this year. He was then, according to his mother's account, attacked with chills, pain in the stomach, vomiting and headache. The latter appeared to be very severe. At the same time the bowels were persistently constipated. His illness continued for seven or eight weeks, and for the greater part of this time he lay moaning with his face buried in the pillow. Finally, his pain seemed to subside, but it was now noticed that he could no longer see, and as he grew stronger and attempted to walk that he staggered and fell, and that, indeed, walking without assistance had become impossible.

His present condition is as follows: When placed upon his feet, he sways badly to and fro, and, if unsupported, falls. He stands with feet widely separated and arms thrown out, as though making a vain effort at balancing. He makes the attempt to walk unwillingly. He staggers badly and tends constantly to fall forward.

An examination of the eyes by Dr. de Schweinitz revealed double optic neuritis with total blindness. Pupils dilated and immovable. No nystagmus.

Hearing and the other special senses appear to be normal. There is no affection of speech. Polyuria, however, is present, and there is general muscular weakness. The knee-jerks are absolutely lost on both sides.

The headache has apparently entirely disappeared. His mother now volunteers the opinion that his head has, of late, been increasing in size, as she has, within a short period, been obliged to purchase for him at short intervals larger hats. The head measures in circumference twenty-one inches (53.2 m.m.).

In the above histories it is to be noted: first, that a double optic neuritis of high grade existed in all four

cases; secondly, that this neuritis was associated with total blindness; and thirdly, that the knee-jerks exhibited the following peculiarities: In Case I they were absent in the early history, but reappeared and were exaggerated later on and at a time when blindness had made its appearance. In Case II blindness and exaggerated knee-jerks were again associated, but in Cases III and IV, though total blindness was present, the knee-jerks were absent.

Before studying these symptoms, let us turn our attention still further to the following group of cases.

CASE V.³—L. H. (Philadelphia Hospital), male; white; family history, so far as could be gathered, negative. In June, 1888, having been exposed to the sun for a long time, he suffered a severe heat-stroke. Recovery from this seems to have been very protracted and never complete. When he left his bed it was noticed that he staggered a great deal as he walked, and had at times headache. At the time of his admission to the Philadelphia Hospital it was noted that his movements were very irregular; they were jerky, and when asked to take hold of an object with his hands, betrayed decided lack of co-ordination. When standing his feet were well separated, the arms extended laterally and his head thrown slightly back. The whole posture suggested the effort of balancing. On attempting to walk it was noticed that he kept his feet widely separated, and that he moved them forward by little, short, jerky steps; that every now and then he stopped, swayed to and fro, balanced himself anew and then started afresh. He occasionally fell, and at one time suffered from a fractured fibula in consequence. His gait was eminently staggering, the tendency to fall forward or backward being very marked. The knee-jerks were diminished, though not absent; there was no nystagmus; his pupils reacted well. The eye-ground failed to reveal an optic neuritis. There was, however, a condition of partial optic atrophy. In addition there was some general mental impairment. Vision, as far as it could be tested, did not seem to be markedly impaired.

Three years after admission this patient died of intercurrent diarrhœa.

³ Partially reported. *International Clinics*, Vol. II., 2d series.

The autopsy revealed, in addition to other changes of minor importance, excessive atrophy and softening of the cerebellum.

CASE VI.—J. K. (Philadelphia Hospital), white, aged 34, female. Family history negative, with the exception that one sister died at eight years of age of sunstroke. Personal history: has always been well until present attack. Has had two healthy children. On July 29, 1892, suffered from heat-stroke, for which she was treated at St. Mary's Hospital. She appeared to have been quite ill as her temperature rose to 112°F. (communicated by St. Mary's Hospital resident); remained unconscious for two days. Was treated repeatedly by ice-bath, and on recovering consciousness tried to speak, but she could be with difficulty understood, as her speech was halting and irregular. Eighteen days later she began to walk, but her gait was very uncertain and she fell to the ground on the slightest provocation. Admitted to the Philadelphia Hospital October 10, 1892. At this time it was noted that she walked very unsteadily; her steps very short and jerky; feet spread wide apart, and there was marked festination, especially on turning around. There were, in addition, jerky movements of the head and arms. No nystagmus; pupils rather large, but equal and respond to light readily. Examination of eye-grounds negative. No impairment of vision. Complained constantly of dull headache. Knee-jerks present, but diminished. Speech halting, jerky and drawling by turns. Replied slowly to tests by the æsthesiometer, but this seemed to be due to the increase in the cerebral reaction time rather than to any diminution of sensation. There was also general motor weakness.

CASE VII.—A. J., aged 38; male; colored (Philadelphia Hospital). Family history negative. Personal history: Had the various diseases of childhood; also variola at thirteen years of age, and at various times malaria. His present trouble began some twelve years ago. Experienced at first formication, or some other obscure sensation in the soles of the feet. Walking seemed to relieve this condition. Some two years later he began, as he thinks, to get "weak in the legs," and one day his knees gave way beneath him and he fell. From that time on he can only walk with the assistance of two canes.

Present condition: On standing with feet close together he at once falls forward. On standing with feet widely separated he sways to and fro. Festination is so great that it is now practically impossible for him to walk. An attempt to do so makes him stagger and fall. There is no muscular weakness. There is in addition marked incoordination in the movements of the arms. Knee-jerks are present and well marked. His speech is dragging and prolonged, so much so that he can with difficulty be understood.

An examination of the eyes by Dr. de Schweinitz, revealed the following: pupillary reaction sluggish but present, constant and vertical, nystagmus of slow movement. When eyes are directed to right or left rapid lateral nystagmus occurs. Upward movement increases vertical nystagmus in each eye. Central vision two-thirds in each eye. Both optic disks gray, and deeper layers slightly reddened on surface, edges clear, veins a little fuller than usual; arteries a little smaller. There is no limitation in the form fields. There is, therefore, nystagmus, imperfect reaction of the pupils to light, and probably an early stage of optic nerve degeneration.

Roughly speaking, in the above three cases blindness and marked eye-ground changes were conspicuous by their absence. The knee-jerks were diminished in Cases V and VI, but were exaggerated in Case VII.

Let us now consider briefly some of the symptoms. The writer is well aware that generalizations based upon a limited number of cases must be accepted with great caution. However, cerebellar physiology is still so obscure that any grouping of associated symptoms cannot fail to be of value.

To begin, let us observe that the optic neuritis when present appears to be of an intense character. Now, it is a matter of common experience that very high grades of neuritis may exist without marked impairment of vision. It would seem, therefore, that the added symptom of blindness gives a special significance to the optic neuritis found in these cases. It would appear that, for some reason, if we have optic neuritis at all in cerebellar disease, that it is apt to be intense in character;

and further, that it is likely to be associated, sooner or later, with total blindness. It seems strange, at first sight, that cerebellar tumors should present a specially high grade neuritis, and more strange still, perhaps, that this neuritis should be accompanied by blindness.

The proximity of the quadrigeminal bodies naturally suggests itself as in some way explaining this blindness. The ataxia and titubation present in these cases refers us, other things equal, to disease of the vermiform process. A consideration of anatomy, as well as the autopsy of Case III, will show that if a growth be situated in the vermiform process, especially anteriorly, and that if this growth continue to enlarge, it will, sooner or later, press upon the superior cerebellar peduncles, and very probably upon the quadrigeminal bodies themselves. Now when we recall the relation which the fibres of the optic tract bear to the primary optic centres, we can readily understand how, if pressure or irritation occur at this point, a neuritis should be a consequence. Further, the irritation being direct, we can, perhaps, understand why the neuritis should be of a high grade; and finally, also, why this neuritis should be associated, sooner or later, with total blindness.

With regard to the deafness which is present in some cases of cerebellar tumor, and which was absolute in Case III, it may, perhaps, be explained in a similar manner. The studies of Spitzka, Monakow, and others, have made it extremely probable that the posterior quadrigeminal bodies stand in the same relation to the auditory fibres as do the anterior to the optic fibres. It would simply be necessary, therefore, to our explanation, that the pressure involve these structures also in order that deafness should be a symptom. This condition of affairs seems to have been actually present in Case III.

Cases I to IV were doubtless all cases of tumor. In Case III this diagnosis was confirmed by autopsy. Cases V, VI and VII are interesting because the absence of blindness coincides with the absence of optic neuritis. Of these cases, at least one, Case V, was demonstrated

by autopsy to be due not to tumor, but to atrophy of the cerebellum.

When we turn our attention to the knee-jerk, we meet with a problem of peculiar difficulty. The tendon jerks stand, in all probability, in definite relation with muscle tonus. Muscle tonus, in turn, is apparently the resultant of a number of complex factors, of which the physiological action of the cerebellum is one. It has been demonstrated by Luciani that among the principal symptoms of ablation of the cerebellum is loss of muscle tonus. It would seem, therefore, that we would be justified in accounting for the loss of knee-jerk when observed in disease of the cerebellum by this loss of muscle tonus. In Case V, it will be remembered, loss of knee-jerk and atrophy of the cerebellum went hand in hand.

Further, it is extremely probable that lesions of the cerebellum act as do lesions elsewhere, in one of two ways; i. e., either by destroying tissue and thus destroying function, or by acting as irritants. It is perhaps in this way that we can account for the fact that in some cases of cerebellar disease the knee-jerk is absent and in others present and exaggerated.

The well-known fact that the knee-jerk may be present or absent at different periods in the history of the same case is illustrated by Case I; in this the knee-jerk, absent at first examination, had reappeared and was exaggerated at the second, ten months later. At the same time total blindness had also set in. Total blindness and exaggerated knee-jerks were also associated in Case II. We might be tempted here into supposing that the pressure forwards of the growth in the vermiform process not only produced the blindness, but also, by a further extension, irritated the motor tracts, and thus influenced the return of the knee-jerks. However, that this explanation is insufficient, or at any rate does not apply to all cases, is illustrated by Cases III and IV, in which total blindness was associated with absent knee-jerk.

